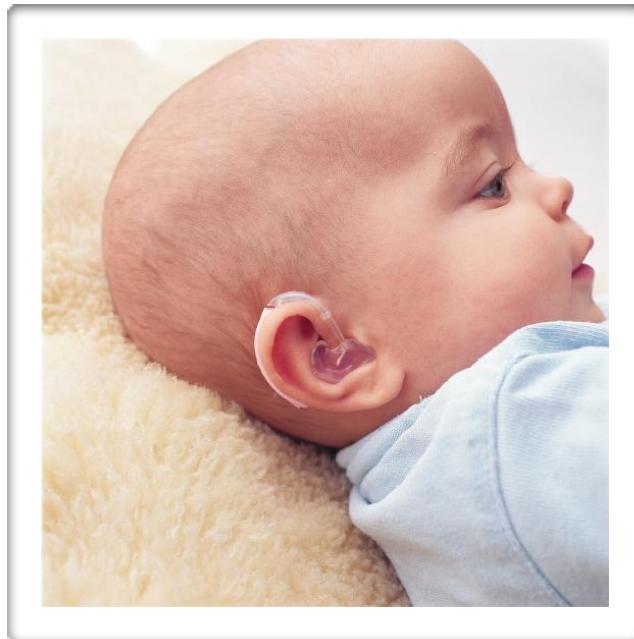


Louisiana Early Hearing Detection and Intervention (LA EHDI) Program



Louisiana Pediatric Diagnostic Audiology Guidelines

Protocols and Standards for Diagnostic Evaluations to Determine Hearing Loss

**Department of Health and Hospitals
Office of Public Health
Hearing, Speech and Vision Services**



These guidelines were developed in part by funds from grants from the Maternal and Child Health Bureau,
and the National Center for Disease Control.

TABLE OF CONTENTS

Introduction.....	3
JCIH Benchmarks and Quality Indicators.....	4
Follow-Up of Failed Hearing Screening.....	5
Evaluation of Newborns and Infants 0-6 Months of Age	6
Evaluation of Children 6 months- 3 years.	9
Use of Sedation in the Evaluation Process	11
Monitoring Children At-Risk for Progressive Hearing Loss.....	12
Amplification.....	13
Reporting Results and Tracking	15
Referral to Early Intervention.....	16
Appendix A: Resources.....	17

INTRODUCTION

The goal of developing this document is to provide guidelines for pediatric diagnostic services that recognize the diversity of individuals and families. These guidelines have been developed specifically for audiological diagnostic services provided to children from **birth-36 months of age**. This is a companion document to the previously created ***Louisiana Hospital Guidelines for Newborn Infant Hearing Screening Programs***.

Audiologists who are not able to provide these services, due to lack of skill, experience, or equipment are ethically obligated by the Louisiana Board of Examiners in Speech Pathology and Audiology Ethics Guidelines to refer families to facilities where the needed services can be obtained.

A well-organized and professional early hearing detection and intervention system can make a significant difference in the lives of children who are deaf or hard of hearing and their families. We hope these guidelines can act as a tool to providing the best services possible.

These guidelines are intended for audiologists who serve infants and young children suspected of having hearing loss. Given the necessity and importance of multi-disciplinary service providers for children and their families, other stakeholders may benefit from these assessment guidelines in the context of early detection and intervention program development.

Who developed these guidelines?

The guidelines on the following pages were developed by a committee of audiologists from the state of Louisiana /Office of Public Health “Sound Start” EHDI Program in collaboration with the Louisiana State Advisory Council on Infant Hearing.

These guidelines are based on the DHH rules and regulations developed to accompany Louisiana Act 653 of 1999 Universal Newborn Screening Legislation and were updated to reflect new recommendations from the Joint Committee on Infant Hearing Position Statement (2007) in November, 2009. See Appendix A.

Funding was provided in part by grants from the US Maternal and Child Health Bureau and Center for Disease Control and Prevention.

JCIH BENCHMARKS AND QUALITY INDICATORS

“The JCIH supports the concept of regular measurements of performance and recommends routine monitoring of these measures for interprogram comparison and continuous quality improvement. Performance benchmarks represent a consensus of expert opinion in the field of newborn hearing screening and intervention. The benchmarks are the minimal requirements that should be attained by high-quality EHDI programs. Frequent measures of quality permit prompt recognition and correction of any unstable component of the EHDI process”. JCIH 2007 Position Statement

Quality Indicators for Screening

- Percentage of all newborn infants who complete screening by 1 month of age; the recommended **benchmark is more than 95%** (age correction for preterm infants is acceptable).
- Percentage of all newborn infants who fail initial screening and fail any subsequent rescreening before comprehensive audiological evaluation; the recommended **benchmark is less than 4%**.

Quality Indicators for Confirmation of Hearing Loss

- Of infants who fail initial screening and any subsequent rescreening, the percentage who complete a comprehensive audiological evaluation by 3 months of age; the recommended **benchmark is 90%**.
- For families who elect amplification, the percentage of infants with confirmed bilateral hearing loss who receive amplification devices within 1 month of confirmation of hearing loss; the recommended **benchmark is 95%**.

Quality Indicators for Early Intervention

- For infants with confirmed hearing loss who qualify for Part C services, the percentage for whom parents have signed an IFSP by no later than 6 months of age; the recommended **benchmark is 90%**.

. The goal for follow up for successful early hearing detection and intervention is referred to as:

1-3-6

- ❖ ***Before 1 month old:***
Complete initial newborn hearing screening
- ❖ ***By 3 months old:***
Complete an appropriate audiological diagnostic assessment on infants failing screening or rescreening
- ❖ ***Before 6 months old:***
Fit amplification and begin early intervention services

FOLLOW-UP ON FAILED HEARING SCREENING

Infants who need further testing due to failed initial hearing screening

- Infants failing the newborn screening are to be referred for follow-up rescreening as soon as possible within two to four weeks from hospital discharge.
- Infants failing the hospital screening must be referred to the primary care physician or to a licensed audiologist for follow-up rescreening. See *Louisiana Audiological Guidelines for Hearing Rescreening of Infants Ver. 3.0, 2013*.

Infants who need further testing due to failed follow-up rescreening

- The audiologist or physician should help the parents to make arrangements for the diagnostic testing at the time of the failed rescreening.
- If possible, diagnostic testing should be performed during the same visit as the rescreening or as soon as possible.
- Only appropriately credentialed and qualified audiologists who possess a valid state license should perform follow-up diagnostic testing.
- **Diagnosis and evaluation of the type and degree of hearing loss should be completed by the time the child is 3 months of age.**

EVALUATION OF NEWBORNS AND INFANTS 0-6 MONTHS OF AGE

Audiological Diagnostic Assessment Protocol

To be considered a diagnostic procedure, ear specific estimates of type, degree, and configuration of the hearing loss must be obtained.

This differs from a simple screening. Adequate confirmation of an infant's hearing status cannot be obtained from a single test measure; rather the initial test battery must include physiologic measures and, if possible, developmentally appropriate behavioral techniques.

1. **Detailed history** should include but is not limited to:
 - a. Parental report of auditory and visual behaviors
 - b. Motor development
 - c. Family history of hearing loss
 - d. History of middle ear pathologies
 - e. Parental concerns
 - f. Prenatal, birth, and neonatal history
 - g. Medical history including: Syndromes or other inheritable conditions, craniofacial anomalies, kidney issues, conditions of limbs/digits, pigmentation issues, exposure to ototoxic medications
2. **Otoscopy**
Visual inspection for obvious structural abnormalities of the pinna and ear canal should be included.
3. **Evoked Otoacoustic Emissions**
Either Transient or Distortion Products Emissions are acceptable.

TEOAE click stimuli: One level (e.g., 80-85 dB SPL) should be completed.
DPOAE stimuli: Use L1/L2 of 65/55 dB SPL.

Pass criterion: An emission of 6 dB signal to noise ratio for at least three frequencies in each ear.

At least one frequency should be located between **2000 and 3000 Hz**
A second frequency should be located between **3000 and 4000 Hz**
The third point could be at any other frequency between **1000 Hz and 6000 Hz**

4. **Acoustic Immitance Testing**

- a. Tympanometry - 660 Hz or higher probe tone
- b. Acoustic Reflex- Ipsilateral middle ear muscle reflex thresholds for 500, 1000, 2000, and 4000 Hz if possible. Currently there is insufficient data for routine use of acoustic middle ear muscle reflex in infants younger than 4 months.

Pass criterion: Type A tympanogram

5. **Diagnostic Auditory Evoked Potential Testing (Non-sedated)**

a. ABR to air-conducted clicks:

Diagnostic testing should include Wave V latency-intensity function responses to at least three differing intensity levels ending with at least one tracing at or below threshold.

Pass Criterion: Normal results consist of Wave V responses for clicks at 25dBnHL within a normal absolute latency range adjusted for the child's corrected gestational age.

b. Change polarity of clicks

Supra-threshold click testing should also include one average with condensation clicks and another average at the same intensity with rarefaction clicks to rule-out **auditory neuropathy/dys-synchrony**.

In a normal ABR the waveforms will be essentially the same morphology and latency with both polarities. If all waveforms in the tracings at one polarity invert when compared to the other polarity, that represents the presence only of the cochlear microphonic (CM) with no neural response. If only the CM is observed, that is consistent with auditory neuropathy/dys-synchrony.

Even though the ABR is abnormal, in this case toneburst testing is not necessary, as it will not yield any additional information.

c. ABR to tonebursts:

In order to obtain more frequency-specific information, ABR stimuli should include at least one low frequency toneburst (such as 500Hz) in combination with clicks.

For even greater specificity both low and high frequency tonebursts could be used in place of clicks (such as 500Hz, 2000Hz and 4000Hz).

If CONDUCTIVE HEARING LOSS is suspected, testing must also include:

c. Bone conduction ABR:

Stimuli should be bone-conducted clicks; masking of the non-test ear should be applied, as appropriate.

Diagnostic testing should include Wave V latency-intensity function responses to at least three differing intensity levels ending with one tracing below threshold.

Diagnostic testing at minimum should include Wave V latency-intensity function responses to at least three differing intensity levels ending with at least one tracing at or below threshold.

Pass Criterion Normal results would consist of Wave V responses at **25dBnHL** for higher frequencies and **35dBnHL** at lower frequencies.

EVALUATION OF INFANTS AND CHILDREN 6 MONTHS TO 3 YEARS OF AGE

Audiological Diagnostic Assessment

1. **Detailed history:** including but not limited to:

- a. Parental report of auditory and visual behaviors
- b. Motor development
- c. Family history of hearing loss
- d. History of middle ear pathologies
- e. Parental concerns
- f. Prenatal, birth, and neonatal history
- g. Medical history including: Syndromes or other inheritable conditions, craniofacial anomalies, kidney issues, conditions of limbs/digits, pigmentation issues, exposure to ototoxic medications

2. **Otoscopy:**

Visual inspection for obvious structural abnormalities of the pinna and ear canal

3. **Evoked Otoacoustic Emissions:** Transients or Distortion Products

TEOAE click stimuli: One level (e.g., 80-85 dB SPL) should be completed.
DPOAE stimuli: Use L1/L2 of 65/55 dB SPL.

Pass criterion: Emission of **6 dB signal to noise ratio for at least three frequencies** in each ear.

At least one frequency should be located between **2000 and 3000 Hz**

A second frequency should be located between **3000 and 4000 Hz**

The third point could be at any other frequency between **1000 Hz and 6000 Hz**

4. **Acoustic Immitance Testing:**

- a. Tympanometry - 660 Hz or higher probe tone in children under 6-18 months, 220 Hz is acceptable in children 18-36 months
- b. Acoustic Reflex- Ipsilateral middle ear muscle reflex thresholds for 500, 1000, 2000, and 4000

Pass Criterion: Type A tympanogram and present acoustic reflexes

5. Behavioral Observation Audiometry (BOA):

In soundfield or with earphones using calibrated stimuli. Insert earphones are recommended if possible.

Pass criterion: minimal and/or startle response at 65 dB.

6. Visual Reinforcement Audiometry (VRA): if appropriate due to child's developmental level.

Stimuli should be speech and also frequency specific tones between 250-6000 Hz. Insert earphones are preferable; sound field may be necessary with some children who will not tolerate earphones.

Pass Criterion: 20 dB to speech and threshold responses at 500, 1000, 2000, and 4000 Hz tones.

7. At least one ABR is recommended as part of a complete audiology diagnostic evaluation for children younger than 3 years old for confirmation of permanent hearing loss.

(The same procedures as outlined in newborn-6 months for recommended ABR procedures apply. See previous section.)

USE OF SEDATION IN THE EVALUATION PROCESS

No child, especially those under the age of six months of age, should be given medication to sedate for testing unless absolutely necessary. Sedating merely for convenience or to speed testing time in a busy clinic schedule is neither ethical audiological practice nor good medical practice. Most normally developing children from birth to 6 months of age can be tested using sleep deprivation and other techniques to induce natural sleep.

The standard is to begin with less medically invasive procedures (i.e. behavioral) and move to more complex procedures (i.e. electrophysiological) requiring or including the use of sedation only when deemed necessary to complete the evaluation.

Children 6 months and older or children with complex medical conditions may need to be sedated to complete necessary diagnostic procedures when **behavioral audiology is inappropriate due to the child's age or other limitations or attempts at behavioral testing have been made** with no success. Conscious sedation is recommended over deep sedation whenever possible.

Administering the sedation and discharging patients after the procedure is not within the scope of practice of the audiologist.

"Moderate Sedation: To gain the cooperation of some infants and young children during physiologic assessments of auditory function, sedation may be required. Yet, sedation of pediatric patients has serious associated risks such as hypoventilation, apnea, airway obstruction, and cardiopulmonary impairment. Consequently, sedative medications should only be administered by or in the presence of individuals skilled in airway management and cardiopulmonary resuscitation. Additionally, the oversight by skilled medical personnel and the availability of age and size-appropriate equipment, medications, and continuous monitoring are essential during procedures and in rescuing the child should an adverse sedation event occur.

The JCAHO has adopted revisions to its anesthesia care standards (JCAHO, 2002), consistent with the standards of the American Society of Anesthesiologists (American Society of Anesthesiologists, 2002). The most current terminology of the American Society of Anesthesiologists has replaced the term *conscious sedation* with *moderate sedation*."

ASHA Guidelines for the Audiologic Assessment of Children from Birth to 5 Years of Age

MONITORING CHILDREN AT-RISK FOR PROGRESSIVE HEARING LOSS

Considerations for managing babies with risk factors for late onset or progressive hearing loss are extremely important. **The timing and number of hearing reevaluations for children with these risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss.**

The JCIH 2007 Position Statement recommends an inclusive strategy of surveillance of **all children** within the medical home on the pediatric periodicity schedule.

All families should receive informational materials that discuss major milestones in normal speech and language development and risk factors for hearing loss in their native language.

Families of infants at high risk should receive additional information on late-onset or progressive hearing loss as well as local diagnostic resource centers.

JCIH Recommendations:

- Infants who pass the neonatal screening but have a risk should have at least 1 diagnostic audiology assessment by **24 to 30 months** of age.
- Early and more frequent assessment may be indicated for children with CMV infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture positive postnatal infections associated with sensorineural hearing loss; for children who have received ECMO or chemotherapy; and when there is caregiver concern or family history of hearing loss.

AMPLIFICATION FOR INFANTS AND CHILDREN BIRTH – 3 YEARS

Most infants and children with bilateral hearing loss and many with unilateral loss benefit from some form of personal amplification.

Pediatric Amplification:

- a. Should definitely be considered for infants and children when the pure tone average or high frequency pure tone average is greater than 25dB HL in at least one ear.
- b. Should be considered when the pure tone average of either ear is greater than 15dB HL and the child is exhibiting speech and language difficulties due to fluctuating or mild hearing loss.
- c. Should be assessed for appropriateness for infants and young children with unilateral loss. Depending on the amount of residual hearing, a hearing aid may be indicated. Contra-lateral routing of signals (CROS) is not currently recommended for children.
- d. Definitive resolution of otitis media (OM) should never delay the fitting of an amplification device. The infant should be referred to the physician for medical management and the audiologist should monitor the status of the OM when determining appropriate prescriptive targets during the hearing aid fitting.

Timeline for amplification fitting:

If the family chooses personal amplification for their child, hearing aid selection should occur **within 1 month of initial confirmation** of hearing loss even if additional audiologic assessment is ongoing.

Amplification/Hearing Aid fitting Guidelines:

The goal of the amplification device fitting is to provide the infant with maximum access to all of the acoustic features of speech within an intensity range that is safe and comfortable. Amplified speech should be comfortably above the infant's sensory threshold but below the level of discomfort across the speech range.

- a. The hearing aid size, make and model should be appropriate for the child's age and development.
- b. For infants under 6 months of age, hearing aid fitting will usually be based on physiologic measures alone using real-ear measurements.
- c. Behavioral threshold assessment using VRA should be obtained **as soon as possible** to cross-check and augment physiologic findings. (JCIH, 2007). Long term monitoring of the validity of the fitting and refinement of the gain and output targets is necessary.

- d. Age appropriate hearing aid prescriptive formulas (such as DSL) that incorporate the use of individual real-ear measurements that account for each infant's ear canal acoustics and hearing loss should be used for fitting infants.

Complementary or alternative technology, such as FM systems or cochlear implants may be recommended as the primary or secondary listening device, depending on the degree of the infant's hearing loss, the goals of auditory habilitation, the infant's acoustic environment, and the family's informed choices.

.

Pediatric Referral for Cochlear Implant:

Cochlear implantation (CI) should be given careful consideration for any child. Since the benefit of CI is variable, a trial fitting of a traditional hearing aid at this time is still recommended by JCIH. The decision to discontinue the hearing aid should be made on the basis of the benefit derived from the amplification.

According to the current FDA guidelines, a cochlear implant:

- a. Is appropriate for children over 12 months of age with profound bilateral sensorineural hearing loss.
- b. May be considered for children 12 months and older with severe bilateral sensorineural hearing loss who are not developing speech and language skills on target after attempting conventional hearing aid use.
- c. May be considered for children 12 months and older diagnosed with auditory neuropathy/dys-synchrony who are not developing speech and language skills on target. A hearing aid trial in patients with AN/AD is still indicated in most cases.
- d. The presence of developmental conditions (e.g., developmental delay, autism) in addition to hearing loss should not preclude the considerations of a CI for an infant or child who is deaf.

REPORTING FOLLOW-UP RESULTS AND LA EHDl TRACKING

REPORTING RESULTS OF ALL RESCREENING AND DIAGNOSTIC TESTING IS EXTREMELY IMPORTANT.

Testing results should be reported on the EHDl **Follow-up Services Report (FSR) Form** which can be obtained by emailing the EHDl Follow-Up Coordinator. This follow-up form should be faxed, or mailed within 7 days to the Department of Health and Hospitals EHDl Program at the address and fax listed on the bottom of the form.

**Email EHDl Follow-Up Coordinator at:
Jeanette.webb@la.gov**

Which results should be reported?

1. Rescreening and/or diagnostic testing results on all **infants who failed a hospital newborn hearing screening even if the results of your testing are normal.**
2. Initial screening or diagnostic testing results on all **infants who never had a hospital newborn hearing screening even if your results are normal.**
3. All results on any child ages birth-5 years of age **identified with a hearing loss for the first time.**
4. All results on any child birth- 5 years of age **fitted with a hearing aid or cochlear implant for the first time.**
5. Every child that is **considered lost to follow-up.** Report information on any child who failed to keep their rescreening or diagnostic testing appointments and who is now considered to be lost to follow-up for your facility.
 - It is recommended that at least two attempts be made to schedule the patient for follow-up testing before reporting them as lost to follow-up. For newborns reported lost to follow-up, by 2 months of age, LA EHDl will use resources to contact the families and encourage follow-up.
 - Communication should be made by at least two different methods before giving up (i.e.: telephone contact and mail contact).
 - Send information on all possible contact family and phone numbers, and information on all previous contacts (phone disconnected, wrong address, other family members, etc).

REFERRAL TO EARLY INTERVENTION

Early intervention is an integral part of the habilitation process for all children identified with hearing loss. Simply establishing an amplification system is only the first step. It is ethically and legally the responsibility of the audiologist to refer all children identified with hearing loss into an early intervention system.

According to Federal Law IDEA, all children diagnosed with a hearing loss **must be referred to Part C (early intervention) within 48 hours.**

In 2009, a new single point of entry into the system of early intervention was developed. The regional Outreach Teachers from PPEP have now been designated as the single point of entry for children identified with any type of hearing loss.

These changes were made in cooperation with Early Steps, DHH EHDI Program, and the PPEP program. These changes assure that families and their children who have hearing loss will have immediate access to:

- an individual with specialized skills, knowledge, and experience in the areas of early childhood language development, hearing loss, communication methods, and family support
- unbiased information on parent and family choices for communication and intervention services as recommended in 2007 Position Statement: Joint Committee on Infant Hearing.

Referrals to regional Outreach Teachers can be made by an audiologist:

**Phone: 225-757-3331; Fax: 225-757-3332;
TDD: 225-769-8160x331; Email: ppep@lalsd.org**

The following information must be provided when referring a child to PPEP:

- Family contact information- at least two phone numbers if possible
- Documentation of hearing loss i.e. written report, audiogram, or copy of the Office of Public Health/Follow-up Services Report

A regional Outreach Teacher will contact the family. Services are provided to families of children who are identified at any age with **any type or degree of either bilateral or unilateral** hearing loss. Additionally, families of infants up to 3 years of age will be referred to Early Steps by the Outreach Teacher in order to receive any additional services the child might need. An additional Early Steps referral from the audiologist is not necessary.

PPEP services are statewide, cost-free, home/community-based services provided to families of children. Services are individualized to meet the needs and priorities of the family. Every family should be referred, and it will be the family's choice to determine their need for services.

Appendix A: RESOURCES

Louisiana Identification of Hearing Impairment in Infants, Acts 1992, No. 417, §1;
Acts 1999, No. 653, §1

http://new.dhh.louisiana.gov/assets/oph/Center-PHCH/Center-PH/hearingspeechvision/LA_LAW.pdf

Rules and Regulations: Chapter 22. Identification of Hearing Impairment in Infants, §2201-2213.

http://new.dhh.louisiana.gov/assets/oph/Center-PHCH/Center-PH/hearingspeechvision/Chapter_22_Rules_and_Regs.pdf

Louisiana Insurance Coverage for Hearing Aids, ACT 816-2003

http://new.dhh.louisiana.gov/assets/oph/Center-PHCH/Center-PH/hearingspeechvision/Hearing_Aid_Legislation.pdf

Joint Committee on Infant Hearing (JCIH) 2007 Position Statement

<http://pediatrics.aappublications.org/content/120/4/898.full?ijkey=oj9BAleq21OIA&keytype=ref&siteid=aapjournals>